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Disclaimer: The opinions or assertions herein are the private views of the authors and are not to be construed as reflecting the views of the Department of the Air Force or the Department of Defense.

INTRODUCTION: Mast cell activation disorders (MCAD) have been associated with Connective Tissue Disorders (CTD) and orthostatic intolerance and/or postural orthostatic tachycardia syndrome (POTS) in limited reports. A single report has also identified kindreds of patients with an autosomal dominant pattern of increased serum tryptase and variably associated CTD, neuropsychiatric findings, atopy, and gastrointestinal diso

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Maj Sofia M. Szari, MD and James M. Quinn, MD
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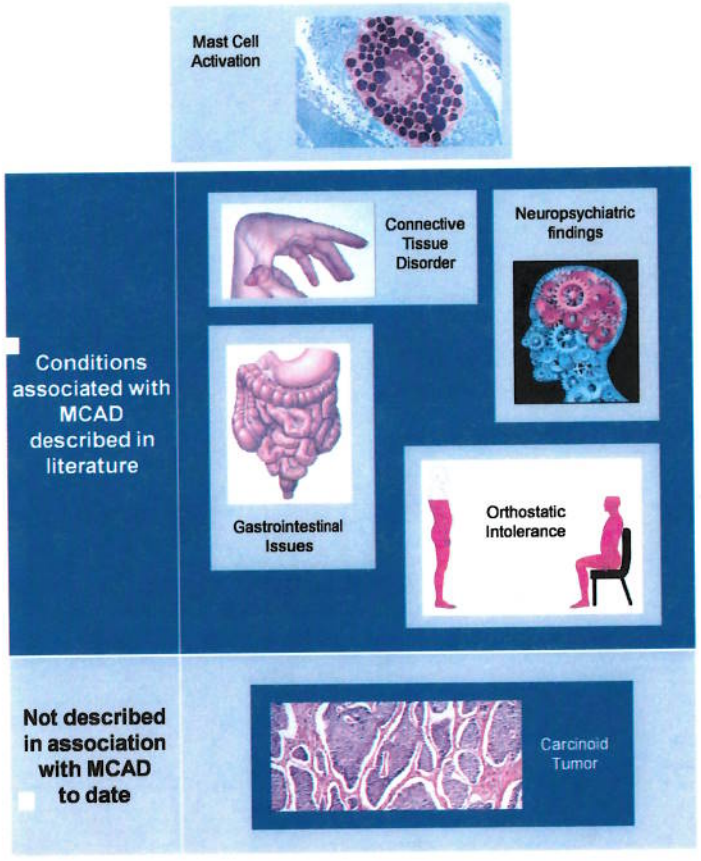
Introduction

Mast cell activation disorders (MCAD) have been associated with Connective Tissue Disorders (CTD) and orthostatic intolerance and/or postural orthostatic tachycardia syndrome (POTS) in limited reports. A single report has also identified kindreds of patients with an autosomal dominant pattern of increased serum tryptase and variably associated CTD, neuropsychiatric findings, atopy, and gastrointestinal disorders. This abstract describes an unusual case of MCAD; Ehlers-Danlos Hypermobility Syndrome (EDHS); orthostatic intolerance; anxiety/ADHD; intermittent diarrhea; and a carcinoid tumor. This cluster of co-morbid conditions with MCAD has not been described to occur in a single patient.

Case Report

A 24 year-old female with a history of fire ant hypersensitivity on immunotherapy, anxiety/ADHD, episodes of orthostatic intolerance, and intermittent diarrhea was found to have an elevated baseline tryptase of 13.7 mcg/L persisting at 13.8 mcg/L on repeat. She was evaluated by hematology/oncology and diagnosed with a MCAD with a low likelihood for systemic disease and offered a bone marrow biopsy. Dermatology consultation concurred with MCAD without evidence of systemic or cutaneous disease. During her evaluation for MCAD, she was diagnosed with EDHS after formal evaluation by geneticist. Shortly after, she underwent appendectomy for acute abdominal pain. Her surgical pathology of the removed appendix revealed a grade 1 carcinoid tumor with negative margins, negative lymph nodes and no inflammation (net G1, path pTaN0).



Discussion

This case illustrates the many potential conditions that can be associated with mast cell disorders and CTD. Connective tissue, namely the components of the extracellular matrix, is known to have affects on mast cells including their migration. It stands to reason that there could be associations with CTD, including orthostatic intolerance and Ehlers-Danlos Syndrome with mast cell disorders. There has been a proposed disease cluster of EDS, POTS and MCAD, which our patient would qualify. She additionally has 2 other associated findings of neuropsychiatric condition (anxiety/ADHD) and gastrointestinal symptoms (intermittent idiopathic diarrhea). Her recent excision of a carcinoid tumor has not been described to date in association with MCAD and is potentially a newly linked association vs incidental.

Conclusion

This case emphasizes the need to evaluate for co-morbid conditions in patients with MCAD. The several conditions associated with mast cell disorders in this patient may represent a newly identified cluster.

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CASE PRESENTATION: A 24 year-old female with a history of fire ant hypersensitivity on immunotherapy, anxiety/ADHD, episodes of orthostatic intolerance, and intermittent diarrhea was found to have an elevated baseline tryptase of 13.7 mcg/L persisting at 13.8 mcg/L on repeat. She was evaluated by hematology/oncology and diagnosed with a MCAD with a low likelihood for systemic disease and offered a bone marrow biopsy. Dermatology consultation concurred with MCAD without evidence of systemic or cutaneous disease. During her evaluation for MCAD, she was diagnosed with EDHS. Shortly after, she underwent appendectomy for acute abdominal pain. Her surgical pathology revealed a grade 1 carcinoid tumor with negative margins, negative lymph nodes and no inflammation (net G1, path pTaN0).

DISCUSSION: This case emphasizes the need to evaluate for co-morbid conditions in patients with MCAD. The several conditions associated with mast cell disorders in this patient may represent a newly identified cluster.

References

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